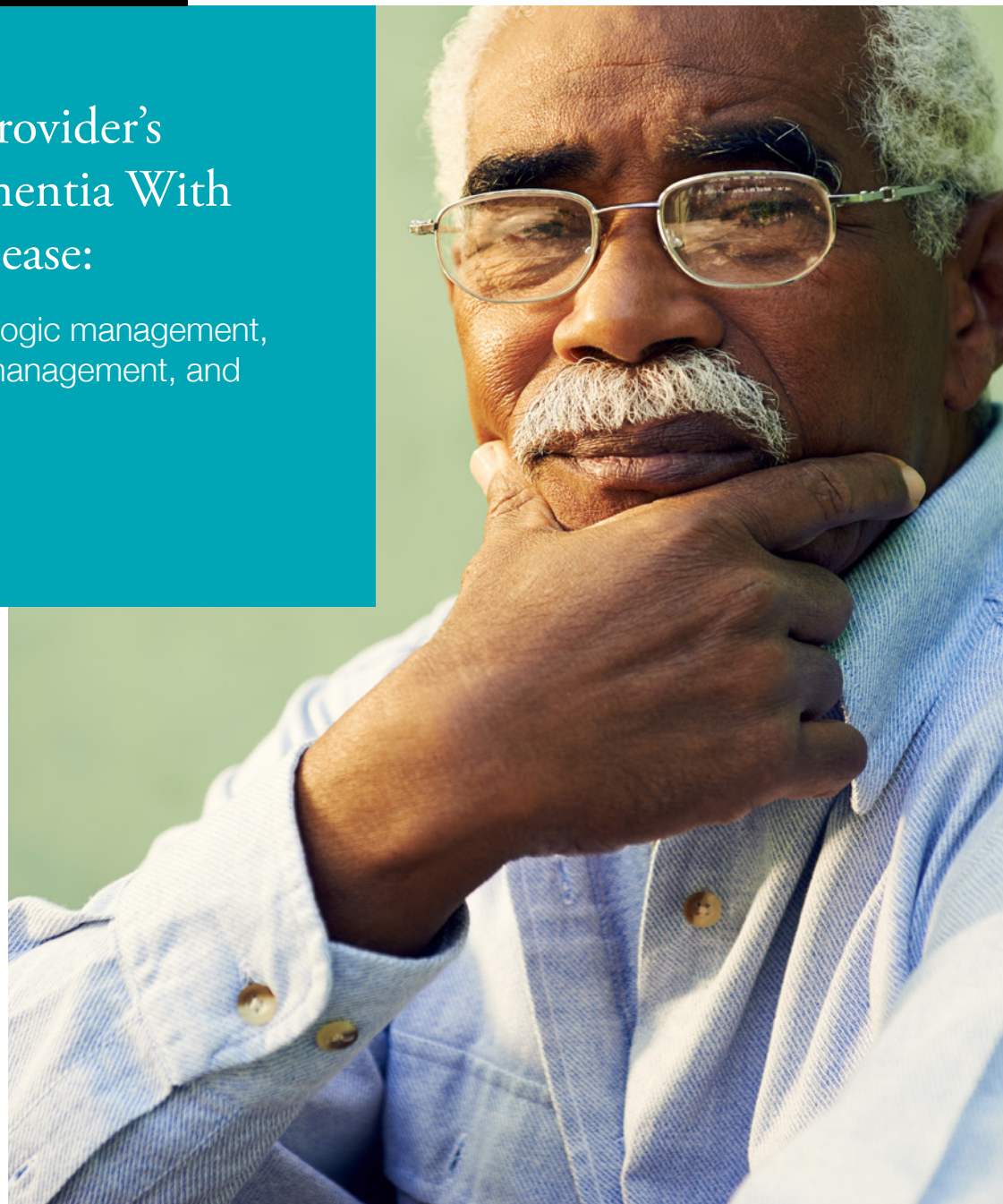


A Healthcare Provider's Guide On Dementia With Lewy Body Disease:

Diagnosis, pharmacologic management,
non-pharmacologic management, and
other considerations

This material is provided
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A Healthcare Provider's Guide On Dementia With Lewy Body Disease: Diagnosis, pharmacologic Management, non-pharmacologic management, and other considerations

Diagnosis

Definition

Dementia with Lewy body disease (DLB) is clinically defined as a progressive decline in cognitive functions, which leads to an impairment of daily function, and which is usually accompanied by some salient features including visual hallucinations, parkinsonism, and cognitive fluctuations.

Etiology

The cause of DLB is unknown. Pathology from autopsy reveals the presence of Lewy bodies on alpha-synuclein immunostaining.

Course

A person with dementia with Lewy bodies may present with progressive decline in memory and visuospatial processing, such as symptoms of short-term memory loss or getting lost while driving. In some cases, there may be a prodrome of dream enactment symptoms, which is suggestive of REM sleep behavior disorder. Some patients may also complain of a longstanding constipation or loss of sense of smell.

Visual hallucinations that are usually well formed and colorful may accompany the symptoms. Patients with DLB may report that they see small animals, people or children and sometimes may have visual misperception where they believe that a tree is a person for example. These hallucinations are often not distressful for the patient. As the disease progresses, these may become more prominent.

One of the core symptoms is the development of parkinsonian symptoms, usually preceding the onset of the cognitive symptoms. Patients with DLB may have masked facies, rigidity, and shuffled gait. The motor symptoms may not be as prominent as in those who have idiopathic Parkinson's disease and in fact

many may not have the typical parkinsonian tremor. As the disease progresses, it may become more difficult for patients to ambulate and other difficulties may arise such as problems with swallowing.

Typically, most patients with DLB have a fluctuating course and their symptoms may appear much more prominent on some days than on others.

Differential Diagnosis

Clinical DLB may appear similar to classical Alzheimer's disease (AD) with some motor features, idiopathic Parkinson's disease or other neurodegenerative entities. It is important to note that DLB and AD can often coexist in the same patient, as evidenced by postmortem studies of patients with DLB. Delirium from systemic illnesses, medications side effects or metabolic abnormalities should be considered, especially if the onset is rapid. Creutzfeldt-Jakob disease could mimic a DLB presentation.

Diagnostic Criteria

The following are the revised criteria for the clinical diagnosis of dementia with Lewy bodies (DLB), as extracted from the third report of the DLB consortium on the diagnosis and management of dementia with Lewy bodies.¹

1. Central feature (essential for a diagnosis of possible or probable DLB)

- Dementia defined as progressive cognitive decline of sufficient magnitude to interfere with normal social or occupational function.
- Prominent or persistent memory impairment may not necessarily occur in the early stages but is usually evident with progression.

- Deficits on tests of attention, executive function, and visuospatial ability may be especially prominent.

2. Core features (two core features are sufficient for a diagnosis of probable DLB, one for possible DLB)

- Fluctuating cognition with pronounced variations in attention and alertness
- Recurrent visual hallucinations that are typically well formed and detailed
- Spontaneous features of parkinsonism

3. Suggestive features (If one or more of these is present in the presence of one or more core features, a diagnosis of probable DLB can be made. In the absence of any core features, one or more suggestive features is sufficient for possible DLB. Probable DLB should not be diagnosed on the basis of suggestive features alone.)

- REM sleep behavior disorder
- Severe neuroleptic sensitivity
- Low dopamine transporter uptake in basal ganglia demonstrated by SPECT or PET imaging

4. Supportive features (commonly present but not proven to have diagnostic specificity)

- Repeated falls and syncope
- Transient, unexplained loss of consciousness
- Severe autonomic dysfunction, e.g., orthostatic hypotension, urinary incontinence
- Hallucinations in other modalities
- Systematized delusions
- Depression
- Relative preservation of medial temporal lobe structures on CT/MRI scan
- Generalized low uptake on SPECT/PET perfusion scan with reduced occipital activity
- Prominent slow wave activity on EEG with temporal lobe transient sharp waves

5. A diagnosis of DLB is less likely

- In the presence of cerebrovascular disease evident as focal neurologic signs or on brain imaging
- In the presence of any other physical illness or brain disorder sufficient to account in part or in total for the clinical picture
- If parkinsonism only appears for the first time at a stage of severe dementia

6. Temporal sequence of symptoms

- DLB should be diagnosed when dementia occurs before or concurrently with parkinsonism (if it is present). The term Parkinson disease dementia (PDD) should be used to describe dementia that occurs in the context of well-established Parkinson disease. In a practice setting the term that is most appropriate to the clinical situation should be used and generic terms such as LB disease are often helpful. In research studies in which distinction needs to be made between DLB and PDD, the existing 1-year rule between the onset of dementia and parkinsonism DLB continues to be recommended. Adoption of other time periods will simply confound data pooling or comparison between studies. In other research settings that may include clinicopathologic studies and clinical trials, both clinical phenotypes may be considered collectively under categories such as LB disease or alpha-synucleinopathy.



Pharmacologic Management

Medications to Use

There is no known cure for DLB. Medications can be used to mitigate the motor, psychiatric and cognitive symptoms in DLB. The pharmacological plan should be designed and implemented carefully as most medications used to treat the motor symptoms of DLB may worsen the neuropsychiatric ones and vice versa.

Cognitive symptoms of DLB may be readily responsive to treatment with cholinesterase inhibitors. Randomized controlled trials in DLB have mostly demonstrated the benefits of rivastigmine (Exelon), however, donepezil can also be initiated.

Parkinsonian symptoms can be treated if necessary with levodopa/carbidopa. The medication should be started at a low

dose and slowly increased, watching closely for any worsening in visual hallucinations or cognitive symptoms.

Visual hallucinations are not always harmful or disruptive, in which case they may not necessarily need pharmacological treatment. That being said, if pharmacological treatment is necessary, cholinesterase inhibitors such as donepezil or rivastigmine can be attempted. Caution is advised as they may worsen the motor symptoms. If doses for cholinesterase inhibitors are maximized and there is still need for further pharmacological treatment, a trial of an antipsychotic could be considered. Although olanzapine and risperidone may be the most efficient at mitigating psychotic symptoms, they are also associated with worsening in parkinsonian symptoms. Therefore, a trial of quetiapine may be wise as a first choice. Again, the doses should be started low and increased gradually, with close monitoring for worsening in motor symptoms.

If symptoms of depression or anxiety are present in people with DLB, selective serotonin reuptake inhibitors (SSRI) may be used as treatment.

Medications to Avoid

Anticholinergic medications (including over-the counter antihistamines such as diphenhydramine) as well as psychotropic medications (especially pain medications such as opioids) should be avoided as they could lead to delirium in patients with DLB, who are already susceptible to psychotropic medications by definition.

Non Pharmacologic Management

Behavioral Approach

It may not be necessary to pharmacologically treat hallucinations in patients with DLB, as they are often harmless and not disruptive. Families can be counseled to recognize the hallucination and distract from it by switching topics. In addition, daily routines can be helpful to help reorient patients with DLB. It is advised that all curtains are closed and lights are turned off at night. Also, going to bed at the same hour every night may be helpful in establishing that routine. During the day, spending time in the daylight, avoiding naps, and engaging in stimulating activities, can be helpful.

Healthy Lifestyle

There are lifestyle habits that promote health and well-being. Research suggests that the combination of good nutrition, physical activity, and mental and social engagement may provide benefit in promoting health although more research is needed to determine the actual mechanisms.^{2,3} A heart-healthy diet (lower in sugar and fat and higher in vegetables and fruit) is considered to be good for both the body and the brain. An example is the Mediterranean diet that promotes nutrition based on fruit, vegetables, nuts, and grains with limits on consumption of red meat and saturated fats. Physical exercise has been associated with improvement of mood and mobility, and a decrease in the risk for falls.^{4,5} Physical activities that are socially engaging (walking or swimming with a friend and participating in exercise groups) can be especially enjoyable. Engagement in activities that are mentally stimulating (crossword puzzles, sudoku, computer games) is encouraged as long as the activity is enjoyable.

The Alzheimer's Association has more information on tips for maintaining your health: [alz.org/we-can-help-brain-health-maintain-your-brain.asp](https://www.alz.org/we-can-help-brain-health-maintain-your-brain.asp)

Sleep

Disrupted sleep can negatively impact memory and thinking, though the mechanisms are not well understood.⁶

Components of sleep hygiene include:

- Avoid napping during the day
- Avoid stimulants such as caffeine, nicotine, and alcohol too close to bedtime
- Get regular exercise
- Avoid eating right before sleep
- Ensure adequate exposure to natural light
- Establish a regular relaxing bedtime routine
- Associate your bed with sleep. It's not a good idea to use your bed to watch TV, listen to the radio, or read.

For more details on sleep hygiene, you can refer to the National Sleep Foundation at [sleepfoundation.org/ask-the-expert/sleep-hygiene](https://www.sleepfoundation.org/ask-the-expert/sleep-hygiene).

Other Considerations

Support Resources

- Lewy Body Dementia Association: [lbda.org](https://www.lbda.org)
- Alzheimer's Association: [alz.org](https://www.alz.org)
- Family Caregiver Alliance: [caregiver.org](https://www.caregiver.org)
- National Institute of Health/National Institute on Aging: [nia.nih.gov/alzheimers](https://www.nia.nih.gov/alzheimers)
- National Institute of Neurological Disorders and Stroke: [ninds.nih.gov/disorders/dementiawithlewybodies/dementiawithlewybodies.htm](https://www.ninds.nih.gov/disorders/dementiawithlewybodies/dementiawithlewybodies.htm)

Research and Clinical Trials

The National Institutes of Health maintains an extensive listing of clinical trials at [clinicaltrials.gov](https://www.clinicaltrials.gov). Academic medical centers may be engaged in research and clinical trials.

Safety

If wandering or getting lost is a concern, refer the patient and family to the MedicAlert+Alzheimer's Association Safe Return program (operated by the Alzheimer's Association) [alz.org/care/dementia-medic-alert-safe-return.asp](https://www.alz.org/care/dementia-medic-alert-safe-return.asp).

Other strategies for ensuring safety concerns may include door alarms and increased supervision.

Driving

Depending on cognitive and motor findings, the patient may be requested to stop driving, complete test of driving abilities through the Department of Motor Vehicles (DMV), or be referred

to a driver's safety course that will assess driving ability. Reporting to the department of motor vehicles should be consistent with state laws. Some states have mandatory reporting requirements: the diagnosis is reported to local health departments who then report to the DMV. Individual state requirements can be found at: dmvusa.com.

Living Situation and Environment

It is important to determine if the patient's residential setting best meets his or her functional and cognitive abilities. Areas of concern may include personal safety (ability to manage medications safely, ability to manage nutritional requirements, ability to manage personal hygiene) and quality of life (activities and engagement that match the person's needs and abilities). Due to social isolation and stigma, many HIV-positive elders live alone and have minimal support. Thus, it is crucial to inquire about living situation, especially in cases of severe dementia where cART adherence may be compromised due to cognitive difficulties.

Types of living situations range from living at home alone or living at home with supervision, to board and care, assisted living, or memory care units.

Elder Abuse

Patients with dementia and their caregivers are vulnerable to abuse. Refer to Adult Protective Services if there is concern for the well-being of the patient or the caregiver.

To locate an APS office in your state, see: napsa-now.org/get-help/help-in-your-area/.

Legal Planning

Provide information about advance directives and durable power of attorney while the patient is in the early stages of disease and able to articulate his or her wishes. Make referrals for legal and financial advice, especially if there are concerns about the patient's judgment, decision-making, or vulnerability. A formal evaluation for capacity may be warranted. The Alzheimer's Association provides a brochure that covers legal planning: alz.org/national/documents/brochure_legalplans.pdf.

- **Advanced Directives**

These documents allow individuals to state their preferences for medical treatments and to select an agent or person to make health care decisions in the event they are unable to do so or if they want someone else to make decisions for them.

- **Power Of Attorney**

A Power of Attorney (POA) is a legal document that gives someone of an individual's choosing the power to act in his or her place. POAs can be for medical or financial matters.

- **Living Will**

A living will is a written, legal document that spells out medical treatments that an individual would and would not want to be used to keep them alive, as well as other decisions such as pain management or organ donation.

Teaching Video for Providers

An example of a physician telling a patient she has dementia: alz.org/health-care-professionals/dementia-diagnosis-diagnostic-tests.asp#alzheimers_diagnosis.



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